Lessons from the Diagnosis and Treatment of Spontaneous Vertebral Arterial Dissection

Case Report

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Summary

A 36-year-old man presented a sudden left occipital headache and right limb weakness after tooth-brushing. Conventional catheter digital subtraction angiography (DSA) showed a left VA occlusion at the crotch of the posterior inferior cerebellar artery. Four days later, the patient got worse. The angiogram showed the left vertebral artery had reopened and the basilar trunk occluded above the AICA. He died two days later and autopsy demonstrated a dissection of the basilar arteries. Based on the autopsy data from the patient in this study, we suggest that the BA dissection might be due to left VA dissection, and placing a stent on the juncture between the uninjured VA and the basilar trunk might be an effective method to prevent fatal BA occlusion.

Introduction

Dissection of the intracranial vertebral artery (VA) has now been recognized as a primary pathogenesis among young and middle-aged patients [1-4]. The most important clinical symptoms of a spontaneous vertebral artery dissection are unilateral headache and neck pain in 55 to 60% [1,2]. In 23% to 43% [2,5], the symptoms are identical to those of Wallenberg syndrome or lateral medullary syndrome (vertigo, ataxia, falling toward the side of the lesion, dysphagia and typical loss of sense of pain and temperature). Generally, it is very serious and greatly affects the clinical course and prognosis for the patients if there is a dissection of the basilar artery (BA), typically occurring via antegrade progression of VA dissection [1-3]. Here we describe a patient with left V4 portion of VA dissection who was diagnosed as cerebral thrombosis and treated with intraarterial thrombolytic therapy but finally died. We also show the rational therapy of the case.

Case Report

A previously healthy 36-year-old man was admitted to our department because of sudden right limb weakness and left occipital headache. Three days before admission, the patient felt a sudden numbness of the right extremities from the right lower limb to the right upper limb while brushing his teeth in the morning; meanwhile, he felt right limb weakness and left occipital headache. He felt nauseous and vomited five times. All the vomits were gastric contents. He presented with alalia and bucking when drinking water, but with no consciousness disturbance, urinary and fecal incontinence or limb spasms. Several hours later, these symptoms worsened. The patient was unable to move his right upper limb and was able to

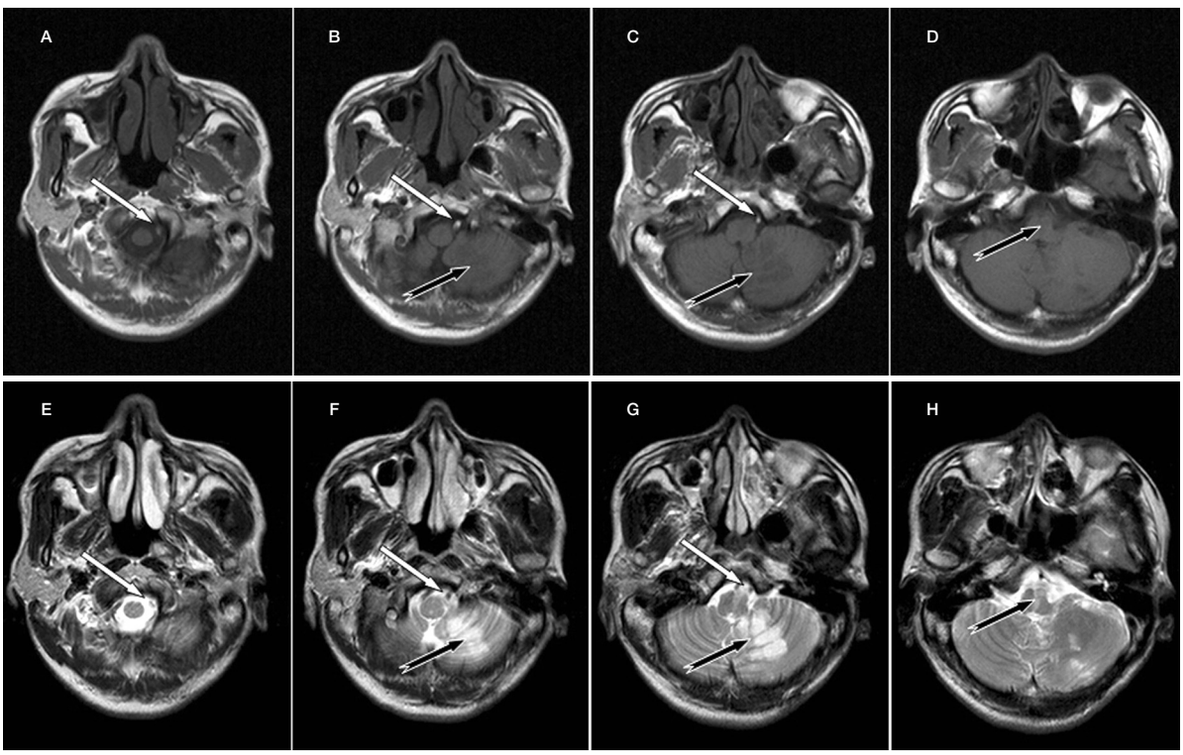

Figure 1 A-H) Axial MR image showing abnormality of left vertebral artery (white arrow) and infarction of left bulb and cerebellum (black arrow). A-D) T1-weighted MR image. E-H) T2-weighted MR image.

move the right lower limb slightly. Cranial CT examination was conducted in another hospital before admission to our department, but no apparent abnormalities were found. The patient was admitted to our department three days after onset of the condition.

History

The patient's right eye was removed because of eye injury in 1985. He had no history of hypertension, diabetes mellitus, hyperlipidemia, heart disease, or migraine. No habit of smoking or alcohol addiction. No family history of stroke.

Examination, diagnosis and treatment

The patient had normal vital signs and cardiopulmonary function. He had no consciousness disturbance. Meningeal irritation sign was negative. He had hyperalgia in the right face and a shallow right nasolabial groove. The mouth seemed tilted leftward. He had mild bucking and dysphagia. The range of motion of the left soft palate diminished and left pharyngeal reflex disappeared. The tongue tilted leftward. The muscle strength of the right limb declined, accompanied by hyperalgia and tendon hyporeflexia. The muscle strength of the right upper limb was assessed to be grade 0 and that of the right lower limb to be grade 2.

Rossolimo sign was positive on the left side, and Babinski and Chaddock signs were positive on the right side. Anhydrosis was noted in the left face. Erythrocyte sedimentation rate, anti-nuclear antigen, electrolytes, complete blood count results, homocysteine, total cholesterol and triglyceride levels were normal. The day after admission, MRI showed an acute infarction in the left cerebellar hemisphere and bulb (Figure 1). Conventional catheter digital subtraction angiography (DSA) on the third day after admission showed a left VA occlusion from the V3V4 junction up to the vertebrobasilar junction (Figure 2).

The diagnosis of cerebellum and bulb infarction was confirmed. Aspirin was administered but the effectiveness was doubtful. At 10:00 am on the seventh day after admission, the patient presented with severe intensive left occipital headache. Local blocking therapy was adopted

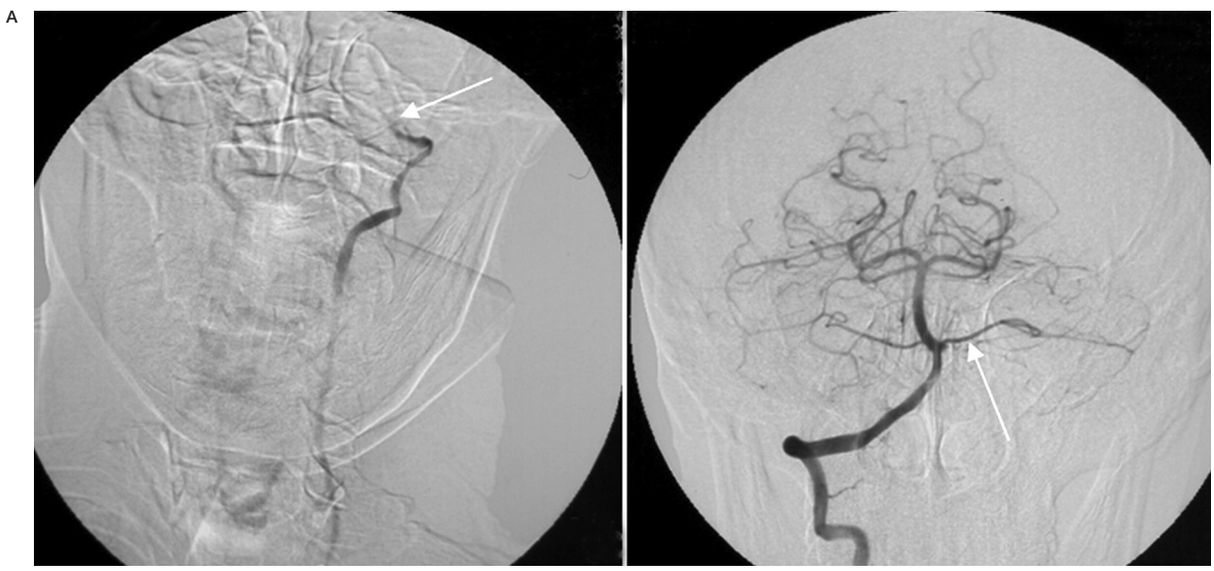

Figure 2 DSA results on day 3 post admission. Left panel, a left VA occlusion at the crotch of the posterior inferior cerebellar artery (arrow). Right panel, the blood supply was partly compensated by the right anterior inferior cerebellar artery (arrow).

but symptoms did not decline, meanwhile dysphasia and right lower limb paralysis worsened. At 2:00 pm the patient could no longer speak, and he suffered from dysphagia and complete paralysis of the right limbs. Repeated MRI exhibited a long T1 and T2 abnormal signals in the left cerebellar hemisphere and medullary bulb, and the lesion was not enlarged significantly when compared with that on admission. At 6:00 pm, the patient presented with consciousness disturbance and vertical and rotatory nystagmus. Corneal reflex disappeared, and pathologic reflexes in bilateral limbs and urinary incontinence appeared. At 7:00 pm, DSA showed recanalization of the intracranial segment of left VA and basilar artery occlusion (Figure 3).

The contrast medium flowed from the left VA to the V4 portion of the right VA. One million IU of urokinase in 100ml normal saline was injected slowly through a microcatheter. Angiograms showed improvement at the middle-lower segment of basilar artery and pontine branches, but occlusion at the basilar artery tip persisted. The microwire and microcatheter could easily pass through the basilar tip to the P2 level of left posterior cerebral artery (PCA), and contrast medium injected with microcatheter showed the P1 level of left PCA was normal. The occlusive sign was seen when the microcatheter was withdrawn to the trunk of basilar artery. The patient's condition progressively worsened and he died on the eighth day after admission.

Autopsy and pathological examination

Autopsy found thrombosis in the intracranial segment of bilateral vertebral arteries, basilar

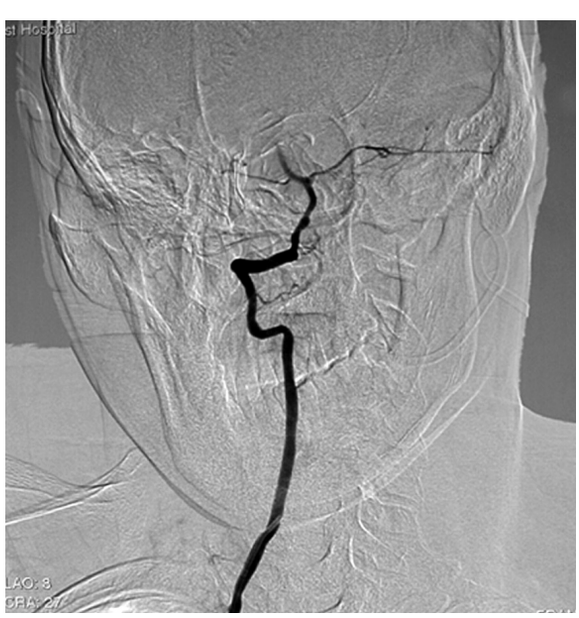

Figure 3 DSA display of BA trunk occlusion after consciousness disturbance of the patient on day 7 post admission.

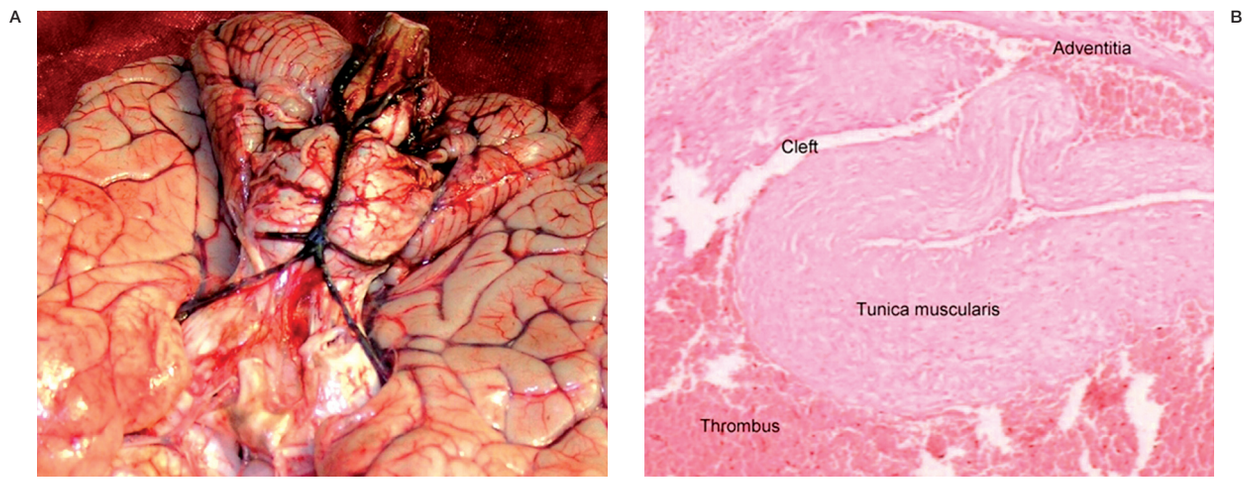

Figure 4 Autopsy and pathologic examination. Left panel, autopsy of the patient, showing the thrombosis in the vertebrobasilar arteries. Right panel, pathologic examination, demonstrating the vascular occlusion and thrombosis between the tunica media and tunica adventitia in the upper segment of BA.

arteries, posterior and superior cerebellar arteries, posterior and anterior inferior cerebellar arteries (Figure 4).

There was localized necrosis in the right vertebral artery, granulation tissue formation in the arterial wall, and the hemorrhagic necrotic foci scattered in the pons, and these lesions were more evident in the left inferior part of the pons. Hypoxic/ischemic changes in brain tissues were noted especially in the occipital and temporal lobes, hippocampus, pons and cerebellum. Pathological examination disclosed the thrombosis was subintimal and compressed the lumen of the upper segment of BA and P1 segment of left PCA (Figure 4).

Discussion

Arterial dissection is a vascular pathological change resulting from arterial intimal damage and blood leakage in between the intima and tunica media or between the tunica media and tunica adventitia after the rupture of vascular endothelium.

The incidence of cervical and vertebral arterial dissections is about 2.5-3/10,000/year and the incidence of VA dissections is about 1.5/10,000/year[6,7]. Ischemic stroke caused by carotid or vertebral arterial dissection accounts for 2% of all cases of ischemic stroke, and 10%-25% of ischemic stroke cases in young people[6-8].

Vertebral arterial dissection tends to involve the intracranial segment of the VA because of its larger range of motion and likelihood of being injured by adjacent bony structures such as cervical vertebrae or styloid processes. Hereditary vascular wall defects, vertebral hyperextension, hyperflexion and rotation are factors inducing vertebral arterial dissections. The main clinical manifestations of a vertebral arterial dissection include headache (70%), and/or localized neurologic impairment (64%). Approximately 10% cases are asymptomatic[4]. The initial symptoms in our patient were right limb numbness and weakness, as well as left occipital pain, and they were closely related to tooth-brushing. It is postulated that the action of tooth-brushing was likely to cause cervical overactivity and, thus, led to the onset of the condition.

MRA and angiography are principal measures to diagnose intracranial vertebral arterial dissection[9,10]. MRA has a good sensitivity for vertebral occlusion and MRI with T1 fat sat sequence as a good tool to demonstrate the mural hematoma. The sensitivity and specificity of MRA for detection of disease in the vertebrobasilar system are 88% and 98% respectively. CT angiography has become one of the main measures to diagnose arterial dissection. Its sensitivity can reach 100%, and the positive and negative predictive values are 98.5% and 98%, respectively[11]. Angiography cannot only reveal the morphology, but also the relationship between the PICA and arterial dissection.

The distinctive artery dissection signs include: bi-lumen sign, pearl-and-string sign, flame sign, aneurysm or fusiform dilatation accompanied by stenosis of adjacent tracts. In addition there is an artery occlusion sign in a small number of patients [12]. Angiography in our patient did not show the typical signs of dissection, but the clinical manifestations, deterioration process, intraarterial thrombolytic practice, and pathological examination strongly prompted VA and BA dissection.

We suggest the pathogenesis as the following: the action of tooth-brushing led to hyperextension of his neck, the intima or the media of the left VA wall disrupted, causing an intramural hematoma in V4 portion up to the left AICA, the left PICA occluded and the patient presented with left occipital headache and opposite neurological defects. Seven days later, left VA dissection progressed to the tip of the BA and the patient presented with intensive headache and progressive consciousness disorders. Considering the failure of thrombolysis therapy in this patient, we held that simple vascular occlusion was not an absolute sign to identify cerebral thrombosis or vascular embolism. Some cases of vertebrobasilar arterial dissection, especially in the intracranial vertebrobasilar arterial system, may present with vascular occlusion.

Intracranial vertebrobasilar arterial dissection is probably related to syphilitic arteritis, arteriosclerosis, vascular wall degeneration, amyoplasia, trauma, and neck surgery [4]. This patient did not have the above-mentioned risk factors, but he might have some precipitating factors, such as small wounds, cervical hyperextension or hyperflexion. For young people, medical history inquiry is essential for the diagnosis of cerebral infarction.

We suggest the following factors strongly suggest a probability of vertebral arterial dissection:

1) young patients;
2) lack of evident risk factors of arteriosclerosis;
3) sudden onset of occipital headache associated with ischemic infarction of posterior circulation;
4) definite arterial dissection signs or unexplainable vascular occlusions. Regarding the treatment of VA dissection, thrombolysis therapy should be used cautiously given lack of definite evidence of thrombosis.

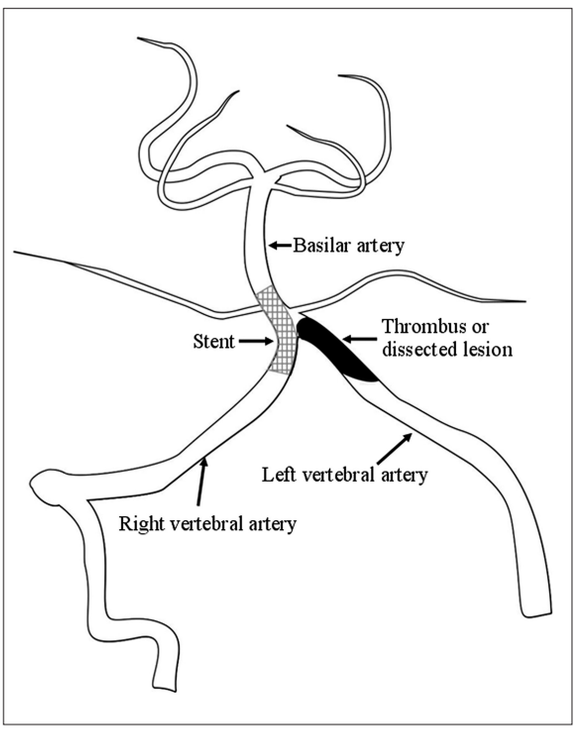

Figure 5 Schematic diagram of stent placement to prevent the progression of thrombus or dissected lesion from vertebral artery to basilar artery.

One side VA occlusion is not fatal, especially under the good condition of the opposite. However, basilar artery occlusion carries a 60% mortality rate and survivors will have a poor quality of life. Therefore, doctors should think highly of this condition when one side VA occlusion occurs near to the basilar trunk. Based on the autopsy data from the patient in this study, provided that there appears aggressive progression of VA dissection to basilar artery, we suggest that placing a stent on the juncture between the uninjured vertebral artery and the basilar trunk might prevent further affections, including thrombus, and basilar artery dissection developed from antegrade progression of VA dissection and thus might become an effective method to prevent fatal basilar artery occlusion (Figure 5).

Acknowledgment

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